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Facial Emotion Identification in Males With Fragile X Syndrome

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Fifteen postpubertal males with fragile X syndrome (FRA(X)) and 15 non-FRA(X) males matched on IQ and age were assessed for their ability to identify the facially expressed emotions of happiness, sadness, anger, fear, disgust, and surprise. Emotions of happiness and sadness were the easiest to identify for both groups of participants. Regardless of etiology, individuals with higher IQ scores performed better at this task than did individuals with lower IQ scores. Results were consistent with findings in females having the fragile X mutation. The current study supported the notion that FRA(X) individuals are sensitive to facial emotion cues presented by others. This finding is discussed in the context of autism and gaze aversion. © 1996 Wiley-Liss, Inc.

KEY WORDS: autism, IQ, mental retardation, sequential processing, social cognition

INTRODUCTION

The ability to discern emotion from facial cues is crucial for normal functioning in society. Deficits in facial emotion identification (FEI) have been detailed in a variety of populations, including right brain-damaged individuals [Borod et al., 1986, 1990, 1993] and people with mental retardation [Gray et al., 1983; McAlpine et al., 1991]. Hobson [1986a,b] has also found that individuals with autism are more impaired in FEI than are individuals with mental retardation matched on mental age. The results of these investigations have characterized FEI as a social cognitive skill heavily dependent on intelligence and mediated in the right hemisphere. George et al. [1993], using positron emission tomography, suggested that the higher order functional neural network for recognizing visually presented facial emotion involves the right anterior cingulate and the bilateral inferior frontal gyri. Happiness and sadness have consistently been the easiest facial emotions to identify, with surprise, fear, disgust, and anger being more difficult.

Fragile X syndrome (FRA(X)) is the most common recognizable form of heritable mental retardation among males [Webb et al., 1986]. Males with FRA(X) have been described as shy and compliant [Chudley, 1984], but also as displaying social behavioral abnormalities such as gaze aversion, aggression, and self-injurious behavior [Bregman et al., 1987; Dykens and Leckman, 1990; Fryns et al., 1984]. Given this behavioral profile, FRA(X) has been associated with autism [Blomquist et al., 1985; Hagerman et al., 1986], although other investigators have disputed this association [Goldfine et al., 1985; Fisch, 1992]. While it is acknowledged that the majority of males with FRA(X) have some autistic features, it is felt that the incidence of true autism [American Psychiatric Association, 1994] in FRA(X) is low.

Mazzocco et al. [1994] compared the facial emotion-matching skills of females with the fragile X mutation to a group of control females. Both unaffected carrier and expressing females with the fragile X mutation were tested. It was found that increasing performance on the matching task was related to increasing IQ but not to fragile X status. A potential confounding variable in the study by Mazzocco et al. [1994] is that the IQ scores of the expressing women were significantly lower than those of the control women. Mazzocco et al. [1994] have called for investigations into the emotion-perception skills of fragile X groups that have been a priori matched on IQ.

Profiles of the cognitive processing abilities of individuals with FRA(X) have also begun to be delineated. Crowe and Hay [1990] administered a variety of right and left hemisphere cognitive measures to individuals with FRA(X) and individuals with Down syndrome. Individuals with FRA(X) performed less well than the Down syndrome group on all 6 of the right hemisphere tasks. Crowe and Hay [1990] concluded that a right hemisphere deficit in cognitive functioning may be a pathognomonic feature of FRA(X). Dykens et al. [1987] utilized the Kaufman Assessment Battery for Children [Kaufman and Kaufman, 1983] to assess sequential and simultaneous processing skills in individuals with

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FRA(X). Dykens et al. [1987] reported relative strengths in FRA(X) males for simultaneous processing tasks, with relative weaknesses for sequential processing tasks. In citing Kaufman et al. [1984], Dykens and Leckman [1990] proposed that FRA(X) males should perform better at interpreting the overall meaning of visual stimuli than at remembering specific details and sequences. In practice, this implies that FRA(X) males should be better at processing pictures, charts, and diagrams than at following a sequence of steps and instructions. FEI thus appears to be a simultaneous, as opposed to a sequential, processing task. FRA(X) males should thus do at least as well on this task as non-FRA(X) males.

The current literature makes divergent predictions regarding the ability of FRA(X) males to identify facial emotion. The right hemisphere deficit profile in FRA(X) males and the possible link with autism predict poorer performance on FEI relative to IQ-matched peers. The simultaneous processing literature and the findings among females with the fragile X mutation predict that FRA(X) males should do at least as well on FEI as IQ-matched individuals. The purpose of the present study is to investigate FEI in males with FRA(X) as compared to a group of individuals with mental retardation matched on IQ.

MATERIALS AND METHODS

Participants

Fifteen postpubertal males with a cytogenetically confirmed diagnosis of FRA(X), ranging in age from 17–66 years ($\bar{x}_{age}$ = 41.80 years, SD = 15.89 years; $\bar{x}_{IQ}$ = 36.60, SD = 7.74), participated in the study. Direct DNA studies confirmed the FMR-1 full mutation in all 12 participants for whom molecular analyses were done; molecular studies were not done on 3 participants. A yoked control group was constructed by matching each FRA(X) individual with a non-FRA(X) individual on the variables of age (±10 years) and IQ (±7 points). This resulted in a control group ranging in age from 16–74 years, with a mean age of 43.53 years (SD = 18.43), and a mean IQ of 37.73 (SD = 9.88). Matched t-tests conducted on age and IQ revealed that the two groups did not significantly differ on these variables. All participants attended one of the educational, vocational, and/or residential programs sponsored by Elwyn, Inc., a large campus-based center for individuals with mental retardation in Southeastern Pennsylvania. IQ scores were gathered from testing routinely performed at this facility under the supervision of the senior author, a licensed psychologist. Individuals with FRA(X) were followed by the Elwyn genetics service, and were still attending Elwyn programs.

Procedure

Forty-eight pages were constructed, with each page having 6 facial photographs in a 2 × 3 array. Each photograph depicted 1 of 6 emotions: happiness, sadness, fear, anger, surprise, or disgust. Photographs were taken from the normed set of facial emotion photographs published by Ekman and Friesen [1975]. Twenty-four of the pages had 6 male models expressing the emotions, and the remaining 24 pages had 6 female models expressing the facial emotions. Photographs were full-face photos from the neck up.

A senior-level psychology student and the first author performed the testing in a quiet area away from other distractions. Participants were seated at a table and were shown a single page of photographs at a time. Testing proceeded using the procedure reported by McAlpine et al. [1991]. This protocol requires little or no overt verbalization on the part of the participant. Each individual was told that he was going to be shown some pictures of people that he did not know. For each page of photographs, a single question was asked; "Show me the person who looks (*emotion*)." Each of the 48 pages was then presented, with a resulting 8 trials for each individual on each of the 6 emotions. Half of the trials for each emotion utilized male models, and half of the trials utilized female models. Participants were given 30 seconds to reply to each question, and their responses were recorded.

RESULTS

All participants were able to complete the entire task within 20 min. Number correct was calculated for each individual at each emotion. In order to assess the effects of fatigue and distractibility on performance in both subject groups, we compared the number of correct answers on the first half of the task to the number correct on the second half. T-tests revealed no significant differences in performance for either subject group. To examine whether stimuli gender differentially affected performance, a t-test compared overall performance on male faces to overall performance on female faces. No significant difference was found. This variable was therefore collapsed in subsequent analyses.

To assess the effect of IQ and its possible interaction with other variables under study, individuals were categorized as either above or below the median IQ of 35.5. This resulted in 15 high-IQ and 15 low-IQ individuals. A 2 (diagnosis) × 2 (IQ) × 6 (emotion) analysis of variance with repeated measures on the third factor was performed on the number-correct scores. This analysis revealed significant main effects of IQ and emotion. No other main effects or interactions were significant. The IQ main effect ($P < .01$, $F_{1,26} = 10.35$) was due to the individuals with higher IQs performing better on the 48 trials than individuals with lower IQs. Table I presents the mean number correct for each group of individuals on the FEI task.

As reflected by the significant emotion main effect ($F_{5,130} = 29.95$, $P < .01$), certain emotions were easier to identify than others: happiness ($\bar{x}$ = 6.00 correct, SD = 1.93), sadness ($\bar{x}$ = 3.00 correct, SD = 2.29), anger ($\bar{x}$ = 2.26 correct, SD = 1.63), disgust ($\bar{x}$ = 2.27 correct, SD = 1.55), surprise ($\bar{x}$ = 1.73 correct, SD = 1.72), and fear ($\bar{x}$

TABLE I. Mean Number Correct and SD (in Parentheses) for Each Group of Individuals

	FRA(X)	Control	Total
High-IQ	20.13 (2.49)	20.74 (2.78)	20.40 (6.94)
Low-IQ	14.29 (1.36)	13.00 (4.00)	13.67 (3.66)
Total	17.40 (1.62)	16.60 (1.77)	17.00 (6.48)

= 1.77 correct, SD = 1.65). Significant orthogonal comparisons ($P < .01$, with 1 and 26 d.f.) revealed that happiness was easier to identify than the 5 other emotions ($F_{sadness} = 51.43$; $F_{anger} = 98.60$; $F_{disgust} = 79.36$; $F_{surprise} = 94.20$; $F_{fear} = 75.43$). Sadness was more easily identified than surprise ($F_{surprise} = 8.98$) and fear ($F_{fear} = 6.89$).

DISCUSSION

The present study provides new information with regard to the social cognitive abilities of postpubertal males with FRA(X), and extends previous work on FEI in individuals with mental retardation. Males with FRA(X) performed as well as age- and IQ-matched controls on an FEI task involving 6 emotions. Furthermore, individuals with FRA(X) showed the same pattern in ease of identification of specific emotions, with happiness and sadness being the easiest to identify. This is a new finding, and does not support the deficit in right hemisphere cognitive processing for males with FRA(X) as detailed by Crowe and Hay [1990]. The FRA(X)-autism link is also not supported by these results, as autistic individuals show a marked deficit in FEI when compared to groups matched for IQ and mental age [Hobson, 1986a, b]. Although individuals with FRA(X) and individuals with autism share some neuroanatomical pathology involving hypoplasia of the cerebellar vermis [Reiss et al., 1991; Courchesne, 1994], the locus of FEI has been identified as the right anterior cingulate and the bilateral inferior frontal gyri. It remains to be seen if future neurological investigations reveal deficits in these areas for autistic individuals.

A large effect of IQ was revealed for both groups of participants. This is in agreement with previous research [Gray et al., 1983; McAlpine et al., 1991], and underscores the large intellectual component of FEI. This finding presents a view of FRA(X) individuals as being able to perform as well as non-FRA(X) individuals with the same IQ level on a socially valid simultaneous processing task. This result supports the work of Dykens et al. [1987] and Dykens and Leckman [1990], and it is also consistent with the findings of Mazzocco et al. [1994] in fragile X females.

Dykens et al. [1987, 1989], however, have also pointed out that individuals with FRA(X) have deficits in the social skills and communication areas of adaptive behavior, but strengths in daily living skills. This profile has been related to a strength in simultaneous processing and a deficit in sequential processing [Dykens and Leckman, 1990]. The finding that FRA(X) individuals do as well on FEI, a skill with a large social component, as IQ-matched non-FRA(X) individuals, tempers the social skills deficit argument. If a social task has a large simultaneous processing component, individuals with FRA(X) may do as well on that task as other individuals of the same intellectual ability. It is important for future researchers to examine specific tasks in the context of the sequential/simultaneous processing model, and not to rely solely on global domains of social, cognitive, or adaptive functioning.

Cohen et al. [1989], using data from the study of parent-child dyads, have postulated that the gaze aversion noted in both individuals with autism and FRA(X) is due to different underlying mechanisms. Gaze aversion in individuals with autism is viewed as the result of an insensitivity to social facial cues. Autistic individuals are said to have an overall dampened sensitivity to another individual's initiating gaze. FRA(X) individuals respond to another's initiation of social gaze by initially averting their gaze, waiting for the individual to look elsewhere, and then returning the gaze. The current study provides evidence that FRA(X) individuals are sensitive to facial emotion cues. FRA(X) individuals also learn the meaning of various facial emotions to the degree expected by their IQ. Further investigations utilizing direct comparison between FRA(X) and autistic individuals are needed to detail the relationship between gaze aversion and facial emotion understanding in these two groups.

The large intellectual component of this and other social tasks also needs to be taken into consideration. Comparison groups used in studies of individuals with FRA(X) need to be matched very closely on IQ or mental age. An equating of means is not sufficient. If individuals are not yoked as in the present study, then differences in etiological groups may not be due to etiology but due to differing IQ variability between groups. Educators must also continue to take intellectual level into account when developing strategies for teaching social skills to individuals with FRA(X).

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